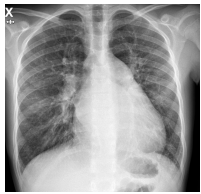
Pulmonary hypertension in Isolated Left Ventricular Apical Hypoplasia (ILVAH) in a pediatric patient

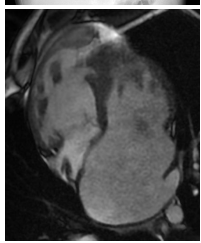
M. Carboni1, F. Ferroni1, G. Agnoletti1, C. Pace Napoleone2
1Pediatric Cardiology, 2Pediatric Cardiosurgery, Regina Margherita Children's Hospital, Torino.

A 12-year-old male was admitted to our hospital after the second episode of syncope in one week. No family history of heart disease nor perinatal problems were reported

.

Lab: NT-proBNP 3700 ng/L.
EKG: enlarged P wave, atrioventricular block type I.
Chest X-ray: signs of pulmonary overload.
Echocardiography: pulmonary hypertension caused by severe mitral regurgitation in a small left ventricle (LV) with a reduced systolic function (ejection fraction 45%).





High doses of diuretics (furosemide and spironolactone) were administered to the patient, who showed a slight clinical improvement but no echocardiographic changes.

Cardiac MRI revealed truncated and spherical LV with abnormal diastolic and systolic function; origin of a complex papillary network in the antero-apical LV; elongated right ventricle (RV) wrapping around the deficient apex. All criterias, except for invagination of fatty material into the myocardium of the defective LV apex, confirmed diagnosis of ILVAH as described by M. Fernandez-Valls et al. for the first time in 2004.

Cardiac catheterization confirmed post-capillary pulmonary hypertension (PCW 22 mmHg, PVRI 4,28 WU/mq).
Endomyocardial biopsy ruled out storage diseases.

Clinical features	Adult patients (n=23)	Pediatric patients (n=10)
Asymptomaticity	2	7
Non-specific symptoms	11	3
Arrhythmias	6 atrial fibrillation 1 atrial flutter 1 ventricular tachycardia	0
Exitus	2	0

A Ventricular Assist Device (Berlin Heart EXCOR) was implanted. One cannula was positioned in the left atrium instead of the LV of reduced dimensions.
Two months later a second cardiac catheterization showed a reduced pulmonary pressure.
Heart transplant was performed without complications.
ILVAH is a rare clinical entity considered asymptomatic during pediatric age. To our knowledge this is the first pediatric case of a ILVAH with pulmonary hypertension that required heart transplant. This case prompt the necessity of an active surveillance to prevent the development of arrhythmias, pulmonary hypertension and bi-ventricular dysfunction in early stage..